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# MYOCLONUS MULTIPLEX AND THE MYOCLONIAS; REPORT OF CASES AND AN ATTEMPT AT CLASSIFICATION.

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Myoclonia is a general term covering all the diseases in which myoclonus or muscle-twitching is a prominent symptom. This is the definition of Foster's Dictionary, and the use of the word in this general sense is the proper one. It has, however, been used by Seeligmüller (1887) to indicate the same thing as paramyoclonus multiplex, and Unverricht used it (1891) in about the same way.

Friedreich reported his case of paramyoclonus multiplex in *Virchow's Archivs*, Vol. 86, 1891. During the ten years following a number of cases which were more or less like his were put on record. When Unverricht published his "Myoclonia" (1891), he recognized that his cases were different from those of Friedreich, and he separated them into a group, called the "familial myoclonias." Following Unverricht, there came reports of a number of cases of myoclonia, some of which resembled this "familial" type, and some, more or less (rather less than more), the original case of Friedreich. The history of these is given in the monograph of Lundborg: "Ueber Degeneration und degenerierte Geschlechter in Schweden," published in 1891, and the more recent monograph of Clark and Prout, *American Journal* *of Insanity*, No. 2, 1902. The criticisms upon these various papers indicated different views. Thus Wollenberg and Boettiger concluded that the Friedreich type of case belonged to hysteria, and that there really was no such independent disorder as paramyoclonus. The "familial" type of Unverricht was thought to belong to the "degenerative chorea" of Huntington. In this view Moebius coincided. Schultz, on the other hand, placed the Friedreich type amongst the forms of convulsive tic and the Unverricht type amongst the degenerative choreas. Marie was of the opinion that paramyoclonus is a special disorder, and with this view Oppenheim, Bechterew and Strümpell appear to agree.

The view which I entertain differs somewhat from any of those expressed heretofore, though it is most in harmony with that of Schultz. It seems to me that the cases which were most in harmony with Friedreich's original case, belong to the group of myokymias, or neuro-muscular spasms without special locomotor effect, and are spasms dependent upon a disturbance of the peripheral motor neurones. There are certainly groups of cases of this kind, as is shown by those recently reported by Hunt[1]. Most of the other cases reported as paramyoclonus belong to the category of hysteria, or functional neuroses, or to convulsive tics. The "familial group" of myoclonias described by Unverricht and Lundborg seem to me to belong certainly to the degenerative choreas, though they are pretty widely separated, as a rule, from the ordinary group of Huntington's chorea, being much more severe in their manifestations, and rapid in their course.

A further discussion of the reasons for this grouping will be given later.

The following cases illustrate four different groups or types of myoclonia.

1. Myoclonia of the spinal and peripheral type, including myokymia, fibrillary myoclonus of Kny, and Friedreich's paramyoclonus multiplex.
2. Myoclonia of functional and hysterical type.
3. Myoclonia of the convulsive tic type.
4. Myoclonia of degenerative chorea and epilepsy. (Myoclonus-epilepsy, myoclonia of family type.)

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[1] Hunt: Journal of Nervous and Mental Disease, June, 1903.

*Personal Cases.* Case I.—*Myokymia and myoclonus (paramyoclonus of Friedreich).* The following case showed very typically the phenomena of myokymia, with at times myoclonia and myotonic contractions with locomotor effect, irregularly distributed and involving muscles not under voluntary control. It is a case resembling in many ways that of Friedreich's, though the dominant feature was the myokymia, and resembles in this more the fibrillary myoclonus of Kny.

*Summary.*—Male, 23, workman, family and personal history negative. At 22 had twitchings of muscles, and painful spasms of legs and arms. Improved, but twitchings never entirely left him; cramps ceased recently. Entered hospital for general weakness, and diffuse pains in bones of legs, pelvis and head. No objective signs of any organic disease. Muscles of all parts of body in continual play of fascicular and fibrillary spasm, causing irregular tremor in fingers and toes, and sometimes locomotor effect of finger and leg. Fascicular spasm in thighs increases and culminates in myoclonus and myotonus of one or both hamstring muscles. Similar myoclonus in biceps. No painful spasm now.

Male, aged twenty-three years, single, Slav, was brought to Bellevue Hospital, complaining of pains in the legs and arms, and great general weakness. He stated that one year ago he had had a severe attack of pains which were called rheumatic. He at that time had also muscular contractions such as were still present and had had at times "cramp-like" contractions of the muscles of the legs and arms. He improved, but continued to have at times cramps, pain, and muscular twitchings. Of late this condition had grown worse. The patient was brought to the hospital, complaining of weakness and pains in the legs, thighs and head. He was a rather small man, somewhat emaciated, but with fairly good muscular development. His body was covered with a fine eruption which was found to be due to scabies.

As the patient lay in bed fine fibrillary spasms could be seen playing over nearly all the muscles of the body. They were very exquisitely marked in the face, where they affected especially the small muscles, such as the buccal, risorius, and nasal. The contractions were not very frequent, not more than one or two being seen at a time, and they occurred in single "flashes," jumping from one spot to another, they affected the two sides about equally. No general motor effect resulted. Similar phenomena occurred in the forearms and in the intrinsic muscles of the hands, the lumbricals and interossei. Here they recurred frequently, causing slight locomotor effect and a condition like a fine tremor, only the fingers were also irregularly flexed, abducted and adducted. When the long flexors were involved the fingers were rhythmically flexed a trifle, the movement sometimes running into a tonus with contraction of the fingers. The movements occurred in the forearms, arms, shoulders, pectoral, abdominal and posterior trunk muscles; here they were more violent, but were less frequent and caused no locomotor effect, except that occasionally the biceps would move the forearm. The calves and intrinsic muscles of the toes were affected just as were the upper extremities. On making the patient lie on his face the movements could be seen most strikingly of all in the muscles of the calf, posterior part of the thigh, and in the buttocks. Here then were frequent fibrillary contractions and also wave-like contractions running along the muscle from center to periphery. On flexing the leg at right angles to the thigh, the inner hamstring muscles would begin to contract more vigorously and finally ended in a distinct myoclonus or more often in a tonic contraction which held the leg flexed for several seconds. The same thing occurred in the outer hamstring muscles and sometimes in both, holding the leg in flexion. Thus there were here fibrillary contractions, wave-contractions, finally ending in a distinct clonic or tonic contraction of the whole muscle, sometimes with locomotor effect. The phenomenon occurred in muscles such as the inner hamstring, not under direct voluntary control. The contractions could be brought out by manipulating the leg, and by blows of the hammer. They were unaccompanied with pain, and the flickering spasms occurred at times 40 to 60 times a minute. The myoclonus and myotonus were observed only when the leg was flexed and the muscles relaxed. A similar condition but much less marked was present in the biceps. The myokymia continued during sleep. There was no atrophy, contracture or paralysis. The general muscular irritability was much increased and immense myoid tremors could everywhere be easily produced. The electric reactions were not tested. The muscular power was fairly good. The deep and skin reflexes were normal. There was no sensory disturbance of any kind. The special senses were normal. He had no stigmata of hysteria. The patient complained of general pains, which seemed to be mainly in the bones. There was no tenderness or swelling of joints or muscles. His mind at first seemed at times confused. He presented no objective symptoms of any visual disorder, or of disease of the central nervous system. The blood and urine were normal, eosinophiles normal. He could walk, but his weakness kept him in bed. He improved and the eruption grew less under mercurial treatment, but the muscular phenomena were unchanged.

Taking the whole history of the case it will be seen that he had painful cramps as in Friedreich's case, clonic twitchings of the muscles of the thighs and arms, not of the peripheral segments, muscular contractions of the whole body of a muscle, sometimes with and sometimes without locomotor effect, and that muscles were affected not under voluntary control. In all these respects, the phenomena were like those of Friedreich's case, but in addition he had a widespread and remarkable myokymia, involving nearly all the muscles. The spasms could not have been of cerebral origin, certainly not cortical, and it seems to me that the seat of the trouble must be either in the spinal cord or peripheral motor neurone,—which is the pathology attributed by Friedreich to his case.

It shows a close relation between ordinary myokymia and the severer types of bundle clonus, to which the term paramyoclonus multiplex, if used at all, should be applied.

Case 2.—*Paroxysmal myoclonus of functional (hysterical and family type. Summary.*—Male, aged forty-two; mother suffered from similar attacks. Healthy till the age of eleven years, when he was kicked in the head by a horse. One month later seizures began, which have lasted thirty-one years, gradually lessening in severity. Seizures come on in groups of short attacks lasting one to five days. Seizures are characterized by some fibrillary tremor, irregular clonic movements, tonic contractions with and without locomotor effect and violent rhythmical movements affecting the face, neck, shoulder and arm muscles and, to some extent, the legs. Muscles of the back are sometimes involved. The muscles involved are symmetrical but not synchronous in their clonic movements.

E. P. B., single, farmer, aged forty-two. Father healthy but of nervous temperament; no phthisis or inebriety, no nervous or mental disease in family. His mother was a healthy woman but during her pregnancy she had attacks of muscular twitchings and jerkings known as "shaking spells." These came on periodically during the day and even interfered with her sleep. They occurred in the latter part of the pregnancy during which she was carrying the patient. After the birth of the child the attacks disappeared. The patient had one healthy sister and lost one sister when she was a baby. He was perfectly healthy until his eleventh year. He was then kicked on the left side of the face by a horse and knocked down, and remained unconscious for three-quarters of an hour. About one month later while sitting quietly in the house he had the first attack of his spasmodic trouble. These attacks would last with intermissions for twenty-four hours, beginning in the night sometimes and lasting till the next night. He would then go for a few days or a week without any, then they would come on again. At times he would have them with short intermissions of a day or two, all one summer, then they would go away for some weeks, then return again. His trouble continued in this way with various remissions till first seen by me, when he was thirty-four years old, at which time he had had the attacks for twenty-three years. During all this time he had no epileptic attacks and nothing resembling epilepsy. He had managed to get his education and attend to his work as a dairyman, spending some time occasionally at sanitariums for treatment.

The year before I saw him he had had an attack of sciatica affecting both legs. This lasted for a month. He had a second attack about four months before I saw him which had lasted all summer. On account of the pains in the legs and the other attacks, he had lost weight, being reduced from 158 to 105 pounds.

When first seen by me at my office he was not having the attacks, and presented the appearance of a nervous, rather anemic and badly nourished man. There was no objective trouble with the muscular system, though the muscles were rather poorly developed. There was some tremor of the hands, rather more in the right, and a little tremor in the legs. The dynamometer showed 30 for right and left hands. There was some fibrillary twitching of the muscles of the back. The skin reflexes were exaggerated. The deep reflexes were normal. There was no disorder in the sensory sphere, the special senses were normal. He had no anesthesia nor ataxia. Bladder and sexual organs were normal.

When the attack came on he was seized with certain rhythmical movements of the head; the hands would shake rhythmically, also the feet, and the lids would quiver. The muscles of the trunk were not involved, but the muscles of the back of the neck would contract and harden, pulling the head back slightly in clonic twitches. When called to see him in one attack at night I found his head moving rapidly antero-posteriorly. Also his arms would be shaken to and fro, but not synchronously, one arm would move more than the other. The trunk and leg muscles did not at that time move very much, though his eyes would blink and his feet and legs would be twitched in irregular clonic movements. He had no pain. The spells would last half a minute, then recur every minute or two, and in this way they would go on for one to five days. They would keep him from sleeping, unless he took some narcotic, then he would go to sleep and the attack would cease. During an attack there was considerable palpitation of the heart, he perspired profusely after the attacks and they would leave him exhausted. Consciousness was never disturbed.

The muscular movements of these attacks, therefore, consisted of violent tremors, tonic contractions of the body muscles without motor effect, clonic contractions irregular in distribution and fibrillary contractions of the body of the muscle which could be observed in between the attacks as well as in the most violent, to precede them. The attacks would occur in short intermissions, every half minute or so, and would last several minutes, and this would go on for from one to five days. These groups of attacks would thus recur every few days, sometimes every week or two, and sometimes intermit for several months.

The patient was not improved by the various measures employed by me, including hyoscine, hypnotics and tonics. He was hypnotized five times, without any special benefit. He was under my observation for one month. He then disappeared, and I heard nothing from him till a short time ago, five years after my first observation, when he wrote me that his general health was excellent and the nervous trouble very much milder, so that when the attacks came on he practically suffered little He states that when they come on he goes to bed, as he is more comfortable there, and passes the time reading until they are over. He had had no epilepsy or hysteria. He is satisfied that it is a family disorder and accepts his affliction philosophically.

Case 3.—*Myoclonia of convulsive tic type. Summary.*—Male, aged forty, no heredity. Abscess of thyroid following typhoid fever; soon after developed clonic, tonic and fibrillary spasms, mainly clonic, affecting first and mostly muscles of abdomen, back and neck; less, those of face, and of forearm; least, those of lower extremities; cease during sleep; no pain; spasms are mostly coördinate, causing grimaces, wry-neck, movements of shoulders and trunk, isolated muscle spasm without locomotor effect, fibrillary and fascicular spasm not marked. Is able to walk and use his hands in delicate work. Spasms vary in intensity without absolute cessation. In four years has gradually improved. No organic disease, general health fair; no epilepsy, mental condition good.

R. M., aged forty years, traveling salesman[1]. The family history is negative and there is in particular no evidence of disease of the nervous or muscular system in the ancestry. Except for the usual diseases of childhood the patient has been well all his life. He denies venereal infections and has been a man of moderate habits. In November, 1896, he had typhoid fever, which kept him in bed for six weeks. The disease ran a mild course without serious complications. During its course, however, a swelling over the thyroid region began to form, which became an abscess. Eight weeks afterwards this abscess broke spontaneously, discharging pus. At the same time the right lower extremity became "awkward and lame," and the patient began to stoop. Shortly afterward he began to have what he called "spasms in the stomach." He was treated with hyoscyamin for this. Slowly the spasmodic discharges began to spread, and by March, 1897, they were very general. In May, 1897, he was treated with thyroid extract, and he says that he was delirious and out of his mind for four weeks. In August, 1897, another tumor of the size of an egg began to form in the thyroid gland, in the location of the previous one. This tumefaction was removed by Dr. Lilienthal, on October 5, 1897. According to a report received from the Mt. Sinai Hospital this tumor was a thyroid cyst. After leaving the hospital the patient says he was well and free from spasms for a period of ten days. They then returned and became continuous, so that he was admitted in August, 1898, to the Montefiore Home, having been ill for over one and a half years. During this time, the spasms, which began in the arms and abdominal muscles, had become generalized. The muscles of the tongue, face, throat and neck had become involved. He would thrust out his tongue and grimace, as in tic, or would throw his head back or twist it to one side, as in wry-neck. He had no pain at any time. Immediately after getting up in the morning the spasms were comparatively slight, but they increased in intensity and reached the climax about four o'clock in the afternoon. Upon going to bed the patient was compelled to take a hypodermic injection of gr. 1-100 hyoscine, and then had a good night's rest. Still at no time of the day, except when he was asleep, did the spasms cease entirely. They were worse when he was excited, and were mildest when he was quietly occupied. He was able to shave himself and to do various kinds of dexterous work with his hands.

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[1] Notes are by Dr. Joseph Fraenkel and myself.

*Status praesens.*—Patient is of medium size, rather poorly nourished, and his skin shows various tattooed figures. He is otherwise free from scars, except a small one over the thyroid region, slightly adherent to the deeper tissues. The skin is of a dirty hue and not very elastic. The muscles are fairly well developed; there are no localized or individual hypertrophies apparent; the muscles are, on the contrary, when not agitated by the spasms, rather flabby and hypotonic. The usual attitude of the patient is a bending forward one, amounting at times almost to a complete doubling up of the body. The head is bent forward to the left and the chin turned to the right. When walking the attitude is the same. He seems to be drawn to the left and shows a sort of left-sided latero-pulsion. It is very difficult for him to assume the erect posture on account of spasms of the abdominal muscles, and attempts increase the spasms.

Temperature 99½ (by mouth); pulse between 130 and 150; pulse is soft, volume rather small; respiration varies from 12 to 18, the rhythm and vigor being constantly interfered with by the spasms.

The thoracic and abdominal viscera are normal.

When the patient is observed, while sitting quietly on the chair, there is a ceaseless and varying play of muscular spasms. These spasms both affect individual muscles and physiologically connected muscle groups, and are essentially clonic, running often into a tonus. The muscles of the trunk, neck and face are affected most. The contractions of the extremities are very much less than those of the rest of the body and the upper extremities show more violent and frequent spasms than the lower; in fact, the lower extremities are, but for the occasional clonic spasms in the quadriceps, almost entirely free; in the upper extremities the muscles nearest the shoulder joint are affected most. The muscles of deglutition and mastication are also affected. There is frequent grinding of the teeth, and swallowing is sometimes interfered with, morsels dropping out of the mouth and fluid passing through the nostrils. There are at times noisy and irregular respirations, due to spasms of the diaphragm. After a number of these severe spasms the patient is covered with perspiration and is considerably exhausted. The frequency of the spasms varies from 10 to 60 per minute.

In the psychical sphere it is noteworthy that the patient cries easily; otherwise no defects are apparent.

Cranial nerves, special senses, gross motor power and cutaneous sensibility are absolutely undisturbed.

Pupillary and skin reflexes are good; tendon reflexes of moderate intensity and easily demonstrative when free from spasmodic contraction. Rectal and vesical sphincters are normal; the patient states, however, that he is unable to urinate except when he is sitting down, and that the spasms cease entirely during defecation and micturition.

The sexual functions are normal. The electrical excitability of muscles and nerves is rather increased, the direct stimulation of the muscles is easily exhaustible. The mechanical irritability of muscles is slightly increased.

A *second examination* was made three months later. There is now a persistent tonic spasm of the abdominal muscles which holds the patient in a stooping position. The recti muscles are thus contracted nearly all the time. They relax only during sleep and for short periods during the day. The pectoral muscles show frequent, bilateral clonic twitchings, at times at the rate of about 50 a minute. Choreic, irregular, not bilateral twitchings are seen from time to time in the muscles of the arms; there is here and there a twitching of the biceps and triceps muscles alone, particularly on the left side. The platysma myoides shows clonic twitchings on the right or left side, but not simultaneously. The right sterno-cleido-mastoid muscle and upper trapezius undergo clonic twitchings and tonic spasm, pulling the head to the left, as in wry-neck. Similar spasms of the left sterno-cleido-mastoid muscle frequently occur, turning the head to the right.

On the whole, the left spinal accessory nerve is more affected than the right, and the spasms are more tonic on this side. Clonic tonic bilateral facial spasms producing prolonged (i.e., for two seconds) grimace occur, and along with them there occur irregular clonic contractions involving the tongue and face muscles.

The patient used to grind the teeth when asleep, but now there is a continual grinding of the teeth and smacking of the lips, as in forms of tic and chorea. There is no nystagmus or disturbance of ocular muscles.

The gluteal muscles go from a state of wavy, fibrillary twitching into a tonic spasm when the patient is put on his back (See Case 1), while the irregular choreic contractions of the lower extremities occur at the same time. To mechanical irritation most of the muscles respond with myoid tremor.

*Third examination,* June, 1900, three and one-half years after the onset of the disease:

The patient's condition is somewhat improved. Still he is in almost constant movement. The spasms continue to affect chiefly the musculature of the neck, shoulders and upper arms, and trunk, the lower extremities being comparatively free; the deep muscles of the back are not affected as much as at the last examination. The number of convulsions per minute is variable, the finer fibrillary contractions occurring as high as forty; the coarser grosser movements occur as high as ten per minute.

The spasms are mostly clonic in character, but also fibrillary and tonic. These movements result in the flexion and extension of the forearms, abduction and elevation of the arms, a flexion of head on thorax, and very slight flexion of trunk on thighs. The peripheral portions of all four extremities are least affected, the movements being greatest in the upper thoracic portions of the body and neck. The diaphragm is still irregularly affected, causing irregular breathing, and to some extent the speech, patient breathing and speaking in intervals. The movements are to some extent symmetrical as to muscles involved, yet not as to time of involvement. Contractions occur irregularly, affect the biceps, then jump to fingers, then to head, then to forearms. They affect usually coördinate groups of muscles, but also individual muscles. Fibrillary twitchings, involving very small limited portions of muscle, and twitching of distinct muscle bundles are seen, the rest of the muscle being apparently at rest. The movements of patient are well nigh continuous under observation, though when unobserved distinct intervals of quiet are apparent, though even then fibrillary contractions are evident.

Effect of position upon movements.—Recumbent posture diminishes movements of trunk, but to a less extent other movements.

Standing increases markedly movements of trunk and somewhat the movements of legs.

Noteworthy that muscles most concerned in standing are least affected, but upper extremities are threshed wildly about.

Standing on one foot possible for a length of time, equal to that possible to a normal person.

Standing with eyes closed, feet together is normal.

Fixation of attention steadies the spasm.

Volitional movements steady spasm, patient being able to shave himself and to do delicate manual work. Things do not drop from hands.

Irritation of skin.—Tickling, pinching, sticking, pressure and cold increase the twitching.

Emotion has a decided staying influence, he asserts.

Alcohol has no effect. Sleep inhibits the movements entirely. Spasms never rhythmical. Tonic spasms are apparent in anterior muscles of neck, yet upon closer investigation they consist of many rapidly occurring clonic contractions.

Sensibility.—No paresthesias. Pains.—In right side of neck and lower and upper extremities. Tactile sense, normal. Pupils equal and normal. Ocular movements normal, but affected by general spasms. Facial innervation equal. Pharyngeal reflexes diminished. No reflexes in upper extremities. Musculature fair in volume, no atrophies. Myotatic irritability increased. Myoidism excessive.

Abdominal reflexes lively. Cremasteric present. Knee-jerks markedly diminished. Achilles ditto. Plantar reflex present;

flexor response. Gross motility normal. No ataxia. Electrical examination: normal reaction.

Pulse 108, full. Respiration variable, 22. Marked prominence of superficial veins of upper extremities and abdomen (caput medusæ). Lungs normal. Heart: no murmurs. Liver apparently enlarged.

*Fourth examination* on March 25, 1902. The patient walks well, but his position is that of extreme kyphosis. There is continual nodding spasm of the head with occasional grimaces of the face. There are no spasms of the eye muscles. The neck muscles are occasionally contracted in tonic spasms which relax at the end of three or four seconds. There are continual rhythmical tonic spasms of the pectoral muscles, drawing the shoulders together, and of the abdominal muscles, pulling the trunk forward. The upper arms are affected in the same way, with occasional tonic spasms involving the whole musculature of the arm, so that there is no actual movement of the forearm. The forearms and hands are not affected by any spasms. There are movements of the back muscles drawing the scapulæ together. He has occasional movements of the intercostals and diaphragm, and owing to this he has, at times, difficulty in breathing. His spasmodic condition, has, on the whole, improved, but he has lost flesh and looks anemic and weak.

At the present time, 1903, seven years since the malady developed, he is much improved, but has still facial grimaces, and neck spasms like those of wry-neck. Mental condition good.

Case 4.—*Myoclonia of type of degenerative chorea, or myoclonus-epilepsy. Summary.*—In 1890, I reported in *Brain*, p. 71, a case of "Chronic Chorea with Autopsy." The patient, a male, aged eighteen years, negative family history, began to suffer with attacks of the chorea of Sydenham at the age of six years. These became chronic and associated later with epileptic attacks. For more than six years the patient suffered from attacks of chorea of a mild type, and occasional nocturnal epilepsy, which did not prevent him from going to school and afterwards to work. The movements increased in violence and extent, and he was finally, at eighteen years, obliged to give up work and go to a hospital. At that time he had not only clonic and choreic movements, but tonic contractions of the muscles of the trunk, fibrillary and wave contractions of the muscles, tremor and not infrequently attacks of an epileptic character. He developed pneumonia and died. I reported the case as one of chronic chorea with epilepsy, but as it appeared in the later stages it would certainly be classed amongst the cases of myoclonus epilepsy, if that classification is adopted.

Case 5.—*Myoclonia of type of degenerative chorea. Summary.*—In 1894, under the title of "The Microbic Origin of Chorea, Case with Autopsy," *American Journal of the Medical Sciences,* January, 1894, I reported a case of a very singular character in a man aged thirty-four years, who had a good family history, who developed rheumatism in the tenth year followed by Sydenham's chorea. This chorea recurred every two or three years, until finally, eight months before he was seen by me, he developed violent choreic and clonic movements, affecting all the muscles of the body, with tonic spasms of the neck and back at times, and rhythmical movements of the head and arms. The case in these last stages resembled Case 2. He subsequently died and an autopsy was made. This man did not have any epileptic attacks, but had a combination of choreic convulsive tic, tonic and rhythmical spasms. The multiple myoclonus, however, was in the last year of his life the dominant feature of the case.

Cases 6, 7, 8.—*Cases of myoclonia of the type of convulsive tic.* In Thompson's "American System of Practical Medicine," under the head of "Choreic Tic" I reported a series of three cases of children who began with what appeared to be ordinary choreic symptoms. In these cases there was first a condition of ordinary choreic movements affecting the face and arms, later tonic and choreic spasmodic movements of the muscles of the neck, causing the patient to appear to have a combination of convulsive tic, or wry-neck, and chorea.

These three cases I called forms of choreiform tic, because beginning with a chorea, like that of the infectious or Sydenham's type, they later developed symptoms of wry-neck and had other tic-like spasms. They belong with the myoclonias of the convulsive tic type, like Case 3.

I have tried to determine the features of myoclonic disorders which ought to be brought out in a description and utilized in classification. Perhaps I have not all, but they seem to be about as follows:

1. The cause.
2. The association with other diseases.
3. The mode of onset.

4. The duration.
5. The persistence or remittance, or intermittance.
6. The groups of muscles or the limbs affected.
7. The symmetry, or bilateral synchronicity.
8. The quickness, or electric character of the spasm.
9. The involvement of muscular groups used coördinately.
10. The involvement of incoördinated groups.
11. The involvement of single muscles not under voluntary control.
12. The motor, or locomotor effect of the twitch.
13. The fibrillary twitching and the twitching of bundles or large masses of the muscle.
14. The predominance of 10, 11, 12 and 13, if several types exist together.
15. The association with tonic or rhythmical spasms, or with epilepsy and mental deterioration.
16. The association with pain, atrophy or paralysis.
17. The condition in sleep, or under anesthesia.
18. The control of the spasm by the will.
19. The influence of emotional excitement on the spasm.
20. The termination.

It is assumed that there is no organic disease and the trouble is not symptomatic of such condition.

Now applying all the tests of character to known and common spasmodic disorders, we find an agreement in the peculiarities of chorea, as it ordinarily develops, i.e., the infectious or Sydenham's type. Nor is there difficulty with symptomatic or degenerative chorea of Huntington.

I think that French and American neurologists also agree pretty well as to what is meant by the different types of spasmodic tic, tic *coordine,* or the tic neurosis of Collins. It includes, to my mind, not only facial tic, but torticollis, and many local tics, as of the diaphragm and larynx, of the trunk muscles, and it may even be generalized as in what I have called progressive or "choreic tic" (myoclonia of the tic type). It is degenerative in origin, and it is a cortical disorder. (I admit cornual or nuclear tics.) It is often a kind of occupation or habit neurosis. It is slow in onset, chronic in character, very nearly continuous in its manifestations. It usually affects the face, neck, the larynx, articulatory muscles, diaphragm or larynx, but it may affect the legs. (I have reported such a case in a boy who had a jumping tic, as well as a speech and facial one.) It is not symmetrical in its distribution. The movements are lightning-like and cause locomotor effects.

There are with it usually no fibrillary twitchings or bundle movements. The clonus may, however, in severe cases, be prolonged into a tonus, and this is the tendency in severe cases Rhythmical tremor and epilepsy may be associated with it. There is no pain, atrophy or paralysis, but rather a tendency to muscular hypertrophy. It ceases during sleep, it can be controlled in a measure by the will, and is made worse by excitement. It is only rarely curable.

The majority of reported cases of paramyoclonus multiplex can be placed in this group as myoclonias of the tic type.

There is another group of myoclonic disorders which has passed under the name of paramyoclonus multiplex.

Its characteristics, if there is such a disease, are claimed to be these:

It is caused oftenest by fright or some psychical disturbance.

Its onset is rather sudden and the duration rather chronic, i.e., months or several years. The spasms intermit, or remit. The muscle groups affected are mainly those of the shoulders, upper arms, thighs and trunk, and the face and periphery are rarely involved. Both sides of the body are affected, but not synchronously.

The movements are lightning-like, and the muscles affected are those of physiological groups, but single muscles and sometimes those not under control of the will may be affected. There is usually decided locomotor effect. There may be tonic and rhythmical spasm (Cases 4 and 5 or group I of Unverricht). The movements cease during sleep and are to a certain extent controlled by the will and increased by emotional excitement.

This type of disorder is distinguished from the tic neuroses by the intermittent duration of the spasms, the involvement of large groups of muscles producing bodily movements and tremors, the absence of spasm of small muscle-groups, causing unilateral facial grimaces and wry-neck movements. The rather favorable course. The neurosis should be placed among the hysterical, or functional groups. My case (Case 2) shows it *may* be a family type and not associated with hysteria.

A fourth group of myoclonias includes myokymia and allied spasms. It is characterized by persistent fibrillary and wave-like contractions of the body of the muscles. Higher and more intense types of this condition show also clonus and tonus of individual muscles. The so-called Friedreich's myoclonus multiplex, the fibrillary myoclonus of Kny would go in this group. They are essentially intrinsic spasms, grosser kinds of fibrillary tremor. So far as I can make out Friedreich's original case was one of this kind, and I agree with Dr. Hunt that they are peripheral or spinal in origin.

Thus I would group the myoclonias under the head of:

1. Myoclonia of Friedreich, or peripheral type, including myokymia.
2. Myoclonia of the functional or hysterical type.
3. Myoclonia of the convulsive tic type, this last type being characterized by many associated spasms, choreic and tonic.
4. Myoclonia of the degenerative chorea or "familial" or myoclonus-epilepsy type, which is closely related with the third.
5. Myoclonia, of the type of the infectious and symptomatic choreas. Although there is sometimes a mixture or fusing of the above groups, they are essentially different disorders, and as a rule can all be easily recognized and easily distinguished from each other. All cases of paramyoclonus multiplex, reported as such, can be grouped in one or the other. Hence there is no need of that much misused term.

*The Myoclonias and their Synonyms.*—The following somewhat interesting list of myoclonias and choreas shows how extensive the nomenclature of muscular spasm has become. I have tried to assign them to the five groups above enumerated. Probably the arrangement is not perfectly correct, or complete, but it is approximately so. I have excluded rhythmical spasms because myoclonia means essentially an irregular spasmodic disorder, a muscle "tumult" (*klonos*), or twitch, or succession of twitches, not an orderly or rhythmical phenomenon like a tremor, though rhythmical spasm may be part of the general picture.

I. Paramyoclonus multiplex, of Friedreich; astasic myoclonia, of Vanlair; multiple spinal myoclonus, of Lowenfeld; fibrillary chorea, of Morvan; fibrillary myoclonus, of Kny.

II. Functional, or hysterical myoclonus multiplex, chorea major, chorea electrica, of Henoch.

III. Myospasmia; memory-spasms, of Friedreich; habit chorea; chorea variable des degenerès; convulsive, or spasmodic tic; tic general; Tourette's disease, myriadrit; palmus; tic neurosis, of Collins.

IV. Degenerative chorea, hereditary chorea, Huntington's chorea; myoclonus-epilepsy; myoclonus, of Unverricht, of "familial" type; myoclonia congenita of Seeligmüller (?); hereditary degenerative chorea of Sachs.

V. Infectious chorea, chorea minor, Sydenham's chorea; chorea electrica of Dubini (?); chorea electrica of Bergeron; senile chorea of Gowers (?).

The stasic myoclonia of Vanlair is apparently a myoid tumor.

As there has been so much discussion over the nature of Friedreich's original case, I append here a translation of the history without adding the author's comments.

## APPENDIX.

### TRANSLATION OF THE ORIGINAL RFPORT OF A CASE OF PARAMYOCLONUS MULTIPLEX. BY PROF. FRIEDREICH.

Ludwig Berlein, fifty years old, carpenter from Mosback (Baden) was taken ill in 1877, with sudden fever, and the appearance of a right-sided pneumonia, following which were developed a chronic induration with shrinking of the upper lobe of the lung, and a dilation of the bronchi of the same lobe. From that time on the patient suffered from dyspnea, chronic cough and mucopurulent expectoration. Lately, also, there appeared progressive emaciation and weakness, which interfered with his work, so that the patient was admitted to the clinic on January 17, 1878. Under the use of liver oil, morphine in the morning, strengthening diet and careful nursing the patient's strength and nourishment improved. Cough and expectoration diminished to a slight remainder, and the patient was able to return to his previous occupation on the 8th of April. But in the fall of the very same year the above-named phenomena returned with renewed violence, so that after a temporary stay in a few other hospitals the patient was for a second time admitted to the clinic on April 29, 1880. The symptoms of bronchitis in the region of the cirrhotic parts of the lung were developed to a high grade, while the lung itself did not expand any.

The left lung was perfectly sound. No fever. No change in the heart or abdominal organs. Two of the sick man's sisters were said to have succumbed to pulmonary tuberculosis.

During the first as well as during the second stay of the patient in the clinic a most peculiar affection of a set of muscles in the upper and lower extremities became noticeable. This showed itself in the form of short rapid contractions, reappearing at short intervals, which affected the biceps and triceps of both arms, and supinator longus of the forearms exclusively.

At the same time chronic spasms showed themselves in the upper part of the thigh involving the vastus externus and internus and especially the rectus femoris, also to a less extent the abductors, and further the biceps and semitendinosus were affected, while the rest of the muscles of the upper part of the thigh were entirely intact. Here and there a muscle of the back or face was slightly affected by this disturbance, which confined itself to that particular muscle exclusively.

Here it was not merely fibrillary or fascicular twitchings but also spasms which involved the whole of the muscle which swelled forward in a mass; and bulged every time, even when the contractions of the muscles were not marked enough to produce a noticeable locomotor effect on the limbs to be moved. Only at times, at an unusually marked contraction, one could notice a slight change in the part to be moved, e. g., during many contractions of the biceps a slight flexion of the forearm or during some of the spasms of the supinator longus a slight supination of the hand.

The spasms of the rectus femoris produced a short and rapid forward movement of the knee joint, that of the biceps and semitendinosus a rapid movement of the part affected, following the tendons which limit the popliteal space.

The spasms, however, were unrhythmical throughout and varying in extent, even though they were separated from each other only by short intervals: (It may be mentioned here, by the way, that by placing the stethoscope on the affected muscles in the moment of their contraction a loud and clear sound, resembling the first heart sound in every respect, could be appreciated.)

Although the affection offered a perfect symmetry in its art, as the very same muscles were affected on both sides, yet every muscle showed a complete independence in relation to its individual contractions. Now it was this muscle, now that muscle, which twitched all for itself without any relation to the others, and if it sometimes happened that the contraction of a muscle on one side took place synchronously with another muscle of the same or other side, or even at times if two symmetrical muscles contracted at the same time, it was only an accidental coincidence, as it may be understood that such an occurrence was made possible by the frequency with which the spasmodic contractions appeared in each single muscle.

The frequency and intensity with which the contractions of each single muscle ensued were not the same on different days or at different times of the day, and in this regard a considerable difference could be noticed. During the times of greater agitation there were 40 to 50 contractions per minute in one and the same muscle, while during the hours of greater rest only 10 to 20 contractions could be counted. Only rarely did it happen that the spasm ceased entirely, or almost entirely, within one-quarter or one-half hour.

Were the spasms of less frequency, so were they also of less intensity, and vice versa. Frequency and intensity always stood in equal relation. It often happened that in one or the other muscle, contractions followed each other very quickly with increasing rapidity, and passed over into a short tetanus lasting about one or two seconds, which was always accompanied by painful sensations in the muscles. In general, the spasms were strongest and most frequent when the patient lay quietly in bed without carrying out any active motions of the extremities. Especially during the evening when the patient attempted to give himself over to the quiet rest of slumber, these cramps in the contracting muscles increased until they produced painful sensations, and thus going to sleep was rendered difficult.

However, if sleep once set in, all the spasms ceased entirely while it lasted. Yet it often happened that the patient was frightened out of his sleep by a sudden very painful, crampy movement of both legs, so that the latter were jerked up against the abdomen. The adjoining individual contractions of the above-named muscles were then always of singular intensity, and it lasted hours until these quieted themselves. But many times, even during the day, although much more rarely than during the night, when the patient lay quietly in bed, there appeared flexion cramps of this sort in the lower extremities, similar to those that are known to appear in cases of acute myelitis.

As soon as one undertook real motions and prolonged active contractions of the affected muscles the spasms immediately disappeared, and the former movements were in no way injured by them.

Thus the patient was not at all disturbed at his work; as soon as the muscles weakened and began to rest, the spasms returned. So during the active extension or flexion of the forearm there were no traces of any spasms in the biceps and triceps of the same side, while the spasm of these muscles of the other quietly lying down continued without weakening.

The lower extremities were in analogous condition, only that in the thigh the painful contractions of the affected muscles continued also during upright position, but here also they entirely disappeared during locomotor actions, as walking. The rough motor power of the affected muscle was in no way affected, and only with the greatest effort could one flex an extended or extend a flexed extremity of the patient against the patient's will.

Station and locomotor coördination were very little affected.

The electric irritability of the affected muscles, as well as those innervated by the same nerve trunk, according to repeated examinations by Prof. Schultze with both kinds of currents, was normal throughout; the mechanical irritability also showed no deviation, and a stroke on the biceps, etc., either by a percussor or with the edge of the hand, showed an idiomuscular swelling in no way higher or more prolonged than one often sees in most healthy individuals. On the other hand, there was an unmistakable increase of reflex irritability of the muscle to skin irritation. Even the effect of cool air on the skin of the exposed extremity increased the frequency and intensity of the spasm. On one occasion during a prolonged exposure, while the patient was being examined, the whole right arm was in painful and violent convulsive tremor, which lasted for one minute. The other skin irritations, such as pinching, pricking, etc., also increased the spasms, and as one would grasp the arm with his hand and squeeze firmly for some time, the spasms of the biceps and triceps would immediately become stronger and more frequent, and often followed each other in such rapid succession that they passed over into a short, lasting tetanus. Here, as it was mentioned above, there could be found no ground for the existence of an increase in the direct mechanical irritability of the muscles, and so this phenomenon was evidently the expression of an increased reflex irritability of the muscles. From the plantar surface one could also bring forth unusually strong reflexes of the lower extremities, and even a slight tickling was sufficient to call forth convulsive tremors of the legs, which repeated themselves after coming to rest a number of times. Tickling of the inner side of the thigh showed marked, although not unusually strong, cremasteric reflexes.

As far as the tendon reflexes were concerned, striking over the tendo-Achillis call forth only very weak contractions of the muscles of the thigh, and no foot phenomena could be brought out, so that in this respect the normal relation existed.

In strong contrast to this there was a marked increase of the patellar reflex; even a slight tap on the tendon of the quadriceps-extensor or ligamentum patellæ was sufficient to produce the most lively and many times repeating extensions of the legs; and when one stretched the tendon of the quadriceps by pulling the patella down, the spasm of the affected muscle appeared, and there ensued painful cramps, which of themselves passed over into an intense tonus, during which time the extremity was strongly extended and the patella drawn forcibly upwards.

On the other hand, irritation and extension of the ligamentum patellæ by pressing it from below upward over the knee joint had no effect, nor were the spasms of quadriceps influenced by it in any way.

All these phenomena which have been described, as well as the muscular spasms and increased skin and patella reflex, were more marked on the right than on the left side. There were no symptoms which would refer to any vasomotor or secretory disturbances. The skin sensations remained normal in every way, and in the same way touch and pain, and differences in temperature were appreciated and localized quickly and precisely. Few phenomena appeared which could be interpreted to mean an injury to the deep muscular sense. There was no atrophy of the affected muscles, excepting the general emaciation and bad nourishment of the whole body, referable to the chronic lung disease.

The physical functions were normal; headache, vertigo, etc., bothered him most. The activity of the higher mental senses was also undisturbed. The pupils were of equal size, somewhat narrow, their reaction to light and accommodation active and normal throughout. There was no fever at any time. Pulse 80 to 90. Some complaints of palpitation. The digestive functions were undisturbed. The secretion and excretion of urine were normal.

The patient attributed the cause of these muscular spasms to a violent fright which he had five years previous at the bursting of a circular saw, and which affected him so thoroughly that he became "purely white" in his face, and at the same time he began to appreciate a sensation of weakness in his whole body; not until fifteen minutes had elapsed was he able to recover himself to any degree. At that accident, however, he received no injury or mechanical shaking up of his body.

About 14 days afterwards the patient noticed these spasms for the first time, and they grew worse later on, but they did not disturb him at his work. Besides that the patient said that he felt the combined sensations of pain and pressure in the arms and legs a number of years before that fright, always, however, only during rest, never during his work or any other active movements.

There was no hereditary predisposition to neurosis.

The patient during his double stay at the clinic was treated with liver oil, with special consideration of his lung disease and in behalf of his improvement of nutrition through which a favorable effect was intended, although the muscular affection remained the same. During the month of March, after only a few galvanizations undertaken by Prof. Schultze, the spasms rapidly began to diminish in intensity, and after a number of days disappeared without leaving a trace.

The increase in the skin and patella reflex diminished in quick succession, so that by the end of April the normal relations were restored in this respect also. The patient was discharged from the clinic by the middle of May.